

Long-term results of complete correction with homograft reconstruction in pulmonary outflow tract atresia¹

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The long-term follow-up over a period of 2 to 5 years of the first 7 survivors of complete correction with homograft reconstruction in pulmonary outflow atresia and extreme Fallot has been studied. Symptomatically, the patients have remained excellent with return to normal of the chest x-rays and electrocardiograms. Investigation has shown continued good function of the homograft aortic valve in the pulmonary position without evidence of progressive pulmonary stenosis or regurgitation. Calcification in the aortic wall is common and occurs quickly but does not interfere with the function of the valve.

In the National Heart Hospital since February 1966, 19 patients have had radical correction for pulmonary atresia and allied conditions, using an aortic homograft to reconstruct the deficient right ventricular outflow tract (Ross and Somerville, 1966). Now that surgical technique for this type of reconstruction is well established the long-term results and durability of the graft are of prime importance. The purpose of this study is to report the detailed follow-up of the first 7 patients who have lived for 2-5 years since operation.

Subjects

Between February 1966 and March 1969, 9 patients had closure of the ventricular septal defect and reconstruction of the pulmonary outflow tract using a stored sterilized aortic homograft. The patients were aged 6 to 34 years. Pulmonary atresia (type 1 or 2) (Somerville, 1970) was present in 6 and extreme Fallot in 3 patients. Relevant details on the 7 survivors are summarized in Table 1. Eight patients had had a previous palliative shunt which was bilateral in 4.

There were 2 deaths. The first death was a woman aged 23 with multiple ventricular septal defects and extreme right outflow and valve hypoplasia, who developed persistent low cardiac output and complete heart block and never left the operating theatre. The other patient aged 14 with type 2 pulmonary atresia died 7 weeks after operation of florid staphylococcal endocarditis on

the patch in the ventricular septal defect which had opened and also caused severe aortic regurgitation. Before this she had had an excellent haemodynamic result but had been kept in hospital for pyrexia and wound infection. The aortic homograft in the pulmonary position was well incorporated in the right ventricle and the suture lines were well healed. The homograft valve cusps looked glistening and 'normal'.

The remaining 7 patients have been examined at 6-monthly intervals, clinically, with chest x-rays in anteroposterior and lateral views, electrocardiograms, and phonocardiograms. Where there was doubt about the presence of calcium in the graft, the patients were screened with an image intensifier.

Six patients were reinvestigated by catheterization, and angiocardiology and serial studies were done in 3 patients.

Surgical technique

As a first step, the aorta is cross-clamped and a short vertical incision is made over the root of the aorta. The Waterston anastomosis is easily identified and is seen to have a firm edge to the ostium. This facilitates closure without tearing and a double layer of 4/0 sutures is used to close the ostium vertically. The aorta is then closed and this part of the procedure should be completed in under 10 minutes of bypass time. Where a preliminary Blalock shunt has been performed, this is secured before bypass is started.

In cases of pulmonary atresia or severe Fallot, our common practice is to incise the right ventricle vertically instead of transversely. This makes it easier to extend the incision up through the area of atresia or valve ring into the vertical incision in the pulmonary artery or region of its

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TABLE I *Clinical data on 7 long-term survivors*

| Case No. | Age (yr) | Diagnosis | Op. date | Before total correction | | Prev. op. | Aortic arch | Complications | Calcif. graft | Pulm. regurg. | State 1971 |
|----------|----------|------------|----------|-------------------------|----------------------------------------|--------------------|-------------|-------------------------------------|---------------|---------------|--------------------------------------------|
| | | | | Hb (g/100 ml) | Resting arterial O ₂ satn % | | | | | | |
| 1 | 9½ | Pulm. atr. | 14.2.66 | 19 | 78 | — | R | Pulm. oedema | + | 1 | Well; pulm. hypertension, ischaemia R. leg |
| 2 | 34 | Fallot | 12.10.66 | 20 | 59 | — | R | — | + | 1 | Normal life |
| 3 | 10 | Pulm. atr. | 17.3.67 | 19.4 | 74-80 | R.Bl. 4 | L | Slight pulm. oedema | + | — | Normal life |
| 4 | 21 | Fallot 1 | 23.2.68 | 21.4 | 61-78 | R.Bl. 2 L.Bl. 5 | L | Transient recurrent laryngeal palsy | + | 1 | R. Blalock open; normal life |
| 5 | 14 | Pulm. atr. | 19.6.68 | 18.4 | 77 | L.Bl. 3 W. 13 | L | — | + | — | Normal life |
| 6 | 11 | Pulm. atr. | 26.6.68 | 20.6 | 47-76 | L.Bl. 3 | L | — | — | 1 | Normal life |
| 7 | 8 | Fallot | 24.3.69 | 16.1 | 90 | L.Bl. 4 | L | R. pleural effusion | — | — | Normal life |

Pulm. atr. = Pulmonary atresia; R. L. Bl. = Right or left Blalock; W. = Waterston anastomosis

bifurcation. Having a vertical incision also means that the reconstructed outflow tract and graft are largely countersunk within the right ventricular outflow making it less liable to compression once the pericardium is closed.

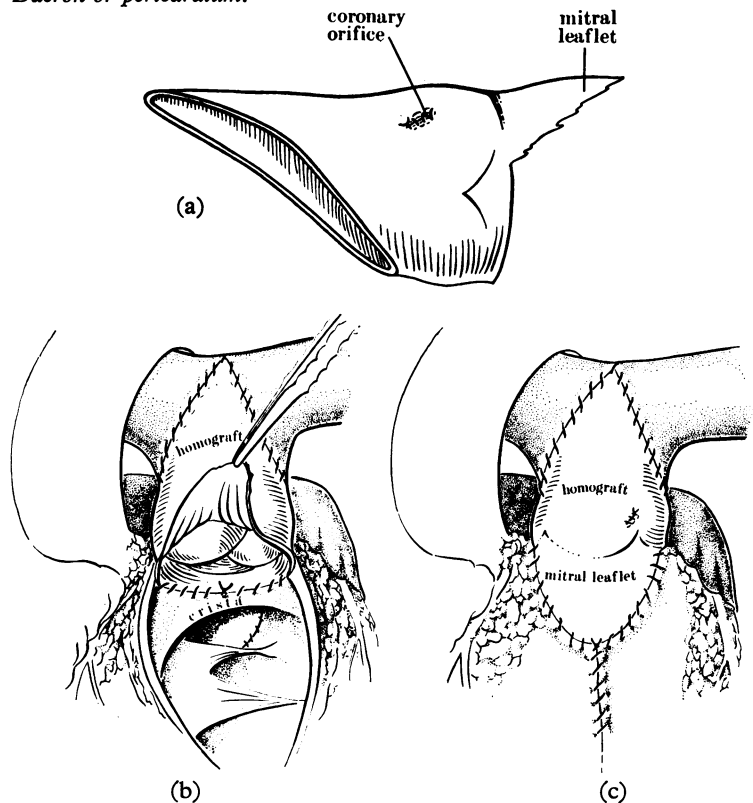
In making a vertical incision caution should be exercised to avoid cutting any anomalous anterior descending coronary artery. In some cases of Fallot with extreme valve ring stenosis, a decision to extend the incision up through the valve ring into the pulmonary artery will be made only after closure of the ventricular septal defect, but it is a safe rule to do a vertical incision in cases of doubt.

Mobilization of the outflow is then carried out by division of trabeculae and the cristae bands rather than by muscle excision at this stage.

The ventricular septal defect is closed first by conventional technique with a patch of knitted Dacron. Sutures are placed through part of the tricuspid valve immediately adjacent to the ventricular septum to avoid damage in the area of the conducting tissue. The remainder of the defect is closed with a continuous stitch.

A homograft aortic valve and aortic segment of adult size, with the attached leaflet of the mitral valve, is chosen. After closing the coronary ostia, the tube and sinuses are rotated so as to bring the anterior mitral leaflet anteriorly. The distal aortic trunk is then bevelled to extend distally towards the pulmonary artery bifurcation (Fig. 1). More aorta is left if the main pulmonary artery is absent as in type 2 pulmonary atresia. The graft is then fixed in place within the right ventricular outflow with a double row of 4/0 sutures, one proximal and one distal. The distal suture line will extend forward to close the incision in the main pulmonary artery if present, or to the bifurcation (Fig. 1b). The proximal suture line fixes the graft to the outflow portion of the right ventricle at the level of the crista supraventricularis and often gains attachment to part of the Dacron septal patch (Fig. 1b). The sutures are continued forward to

FIG. 1 (a) The aortic homograft with bevelled distal aortic trunk and attached anterior mitral leaflet. (b) The distal suture line of the aortic graft extends forward to close the incision in the main pulmonary artery. (c) The anterior mitral leaflet is used to close the right ventriculotomy. This sometimes requires reinforcement with a patch of Dacron or pericardium.



- incorporate the mitral cusp into the adjacent walls of the incised right ventricle. In some cases the mitral valve cusps will be sufficient to close the ventriculotomy, while in others it is necessary to include an additional gusset of pericardium or Dacron (Fig. 1c). In this series being reported, freeze-dried stored aortic homografts were inserted, but fresh homografts are currently used.
- At the completion of the reconstruction the homograft should lie comfortably within the right ventriculotomy.

If an atrial septal defect is present, it is closed at this stage.

Results

- There have been no late deaths and the 7 patients have remained well to the present time, 2½ to 5½ years after the reconstructive procedure.

Complications Case 1 has been well for 5 years and considers he has a normal effort tolerance. He has ischaemic pain in the right leg related to damage done to the femoral

artery by a Seldinger aortogram undertaken after operation to demonstrate persistent large systemic collaterals. He has been advised against strenuous exercise as he has persistent pulmonary hypertension and an aneurysm in the right ventricular outflow tract in the region of the pericardial patch which has not altered in size during the past 3 years and is calcifying.

There have been no complications during the 23 'patient years' survived by the other 6 patients.

Clinical features All patients have remained asymptomatic with normal effort tolerance. None is athletic nor has this inclination.

Accelerated growth rate always occurred during the first postoperative year and Case 4 took 2 years to catch up and equal her normal twin in height.

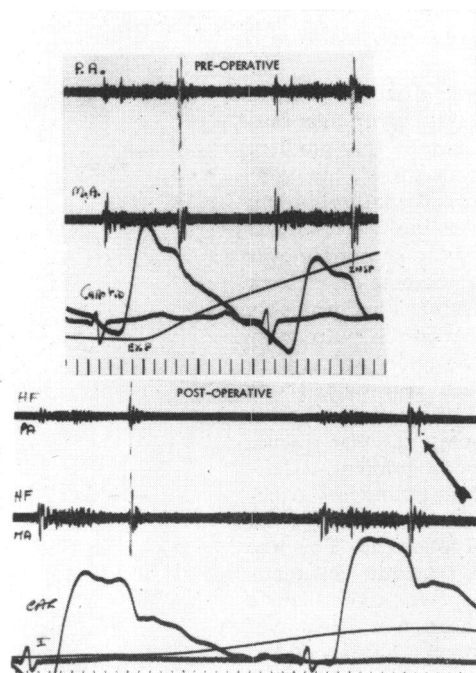
No patient has central cyanosis or clubbing and all have a normal pulse in sinus rhythm. Jugular venous pressure remained high for 3-4 months in 4 patients but afterwards was normal in all but Case 1 in whom it was persistently raised for 3 years and still shows a large A wave at rest. In 2 others, the venous pulsations are unusually high after mild effort, suggesting the compliance of the right ventricle remains abnormal.

Right ventricle pulsation remains excessive in Case 1 where the aneurysm has been palpable since the third post-operative month. The left ventricle is normally palpable in 6 patients. On auscultation, an ejection sound in the pulmonary area is present in 4, and pulmonary valve closure is clear in all and accentuated in Case 1. Movement of the second sound is present in 6 (Fig. 2) but remains widely fixed in Case 1 and not closing in 3 patients, all of whom have complete right bundle-branch block which dates from the operative period. A long pulmonary ejection systolic murmur is present in 3, and 4 have the murmur of pulmonary regurgitation. A soft continuous murmur in the right upper chest persisted after operation in Case 4 and Case 1.

There has been no diminution or increasing delay in P₂ over successive years of follow-up.

Chest x-ray and screening The heart size increased in 5 patients in comparison with the preoperative state, and in 2 who had a large functioning Waterston anastomosis before operation it became smaller 6 months after total correction. The pulmonary artery shadows increased in size and the lung fields appeared to be normally vascularized by the

FIG. 2 Phonocardiogram from Case 3 before and after total correction. In the preoperative record, the second sound is single and there is a systolic ejection sound in the pulmonary area recording. The postoperative phonocardiogram shows a normally moving pulmonary valve closure (P₂) and loss of the ejection sound.



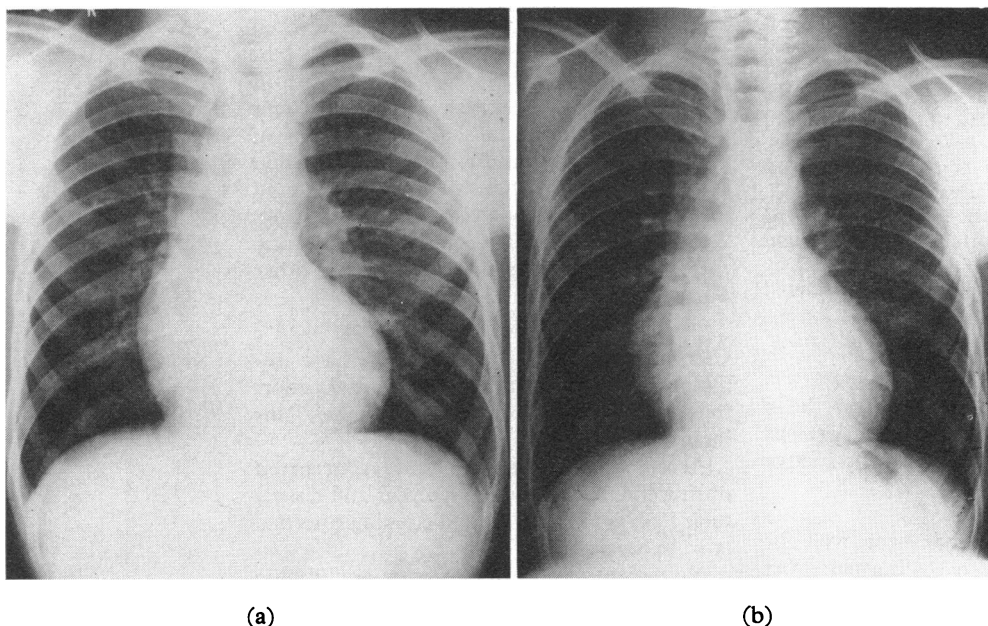


FIG. 3 Chest x-ray from Case 5 before and after total correction. (a) Before correction but 8 years after a left Blalock. (b) 18 months after total correction showing filling in of the pulmonary bay by the enlarged right ventricular outflow and pulmonary root, normal pulmonary vascularity, and a less prominent aortic knuckle.

end of one year (Fig. 3). Calcification appeared in the aortic wall bearing the homograft in 4 by the end of one year, and in a fifth it was visible after 18 months (Fig. 4). In 2 patients (Cases 3 and 7) no calcium was seen on screening or penetrated chest plates 2 and 3 years after operation. Calcification increased to involve the length of the aortic tube over a period of 3 to 4 years but never appeared to involve the valve cusps.

Severe pulmonary oedema occurred in Case 1 and lasted for several weeks. Transient pulmonary oedema occurred in Case 1 and lasted for a few days. Transient pulmonary oedema appeared for the first few days in Case 4 who was extremely cyanosed before operation.

Electrocardiograms Five patients developed complete right bundle-branch block at operation, but in 4 the secondary R wave in one diminished in size over the succeeding 2 to 4 years (Fig. 5), suggesting loss of right ventricular hypertrophy. In Case 1 with persistent right ventricular hypertension, the steep T inversion and large R waves persisted over the right chest leads (Fig. 6). Right ventricular hypertrophy regressed almost completely in the other 2 patients (Fig. 7). No patient had dysrhythmias, but left axis deviation appeared in 2 patients. Left ven-

tricular voltage increased to normal in all patients.

Investigation Haemodynamic data were obtained in 6 patients. One patient (Case 2) refused investigation, stating that 'as there was nothing wrong it could not be necessary'. Three patients had recatheterization 1 to 3 years after the first study.

Peak systolic gradients of 20–50 mmHg were recorded across the right ventricular outflow tract. The major part of this gradient was across the 'pulmonary valve', but 5–15 mmHg was recorded across the infundibulum in 3 patients. Serial studies in Case 1 showed an increased gradient of 25 to 55 mmHg over 4 years as well as an increase in resting pulmonary artery pressure. This probably does not represent an increase in pulmonary vascular resistance as the child was still in severe heart failure when investigated one month after operation. The response to effort suggested severe pulmonary vascular disease but there was no increase in gradient. A left-to-right shunt into the right pulmonary artery was identified in Cases 1 and 4 and persisted over the 4-year period of follow-up. This was due to persistent large systemic collaterals (Case 1, Fig. 10) and a patent right Blalock (Case 4) which escaped ligation at the time of operation. Two other patients (Cases 3 and 6)

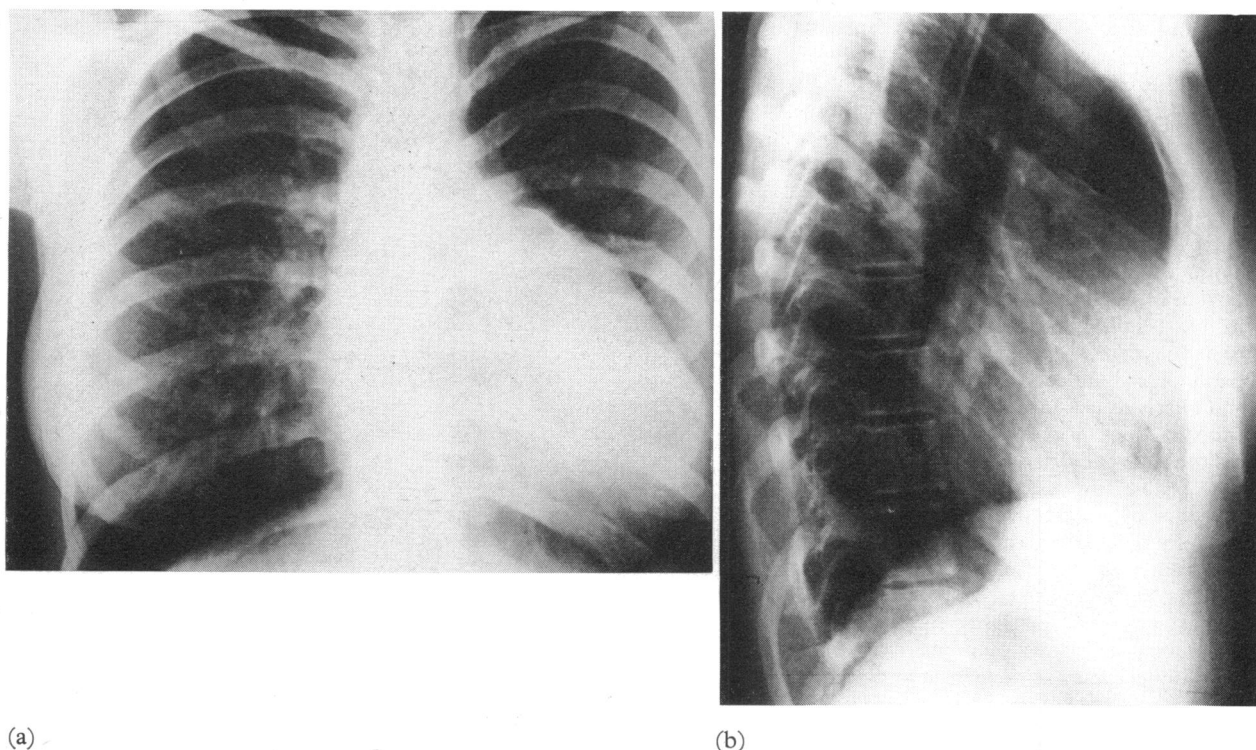


FIG. 4 Calcification in Case 3 in the wall of the aorta bearing the homograft, 4 years after total correction.

showed no increase in gradient across the 'pulmonary' valve in serial studies, and in 4 others there was no change in gradient on mild exercise or rise in the right ventricle and diastolic pressures. Four patients had trivial pulmonary regurgitation which had not changed in those with serial studies. The right ventricle contracted well, and in all patients the graft cusps were moving normally and showed no sign of thickening or calcification. The pulmonary vascular pattern looked near normal in all (Fig. 8) except Case 1 where peripheral appearances suggested severe pulmonary vascular disease. Aortography confirmed the presence of large collateral arteries in Case 1 but was not done routinely in the other patients.

Discussion

Now that the operative technique has been standardized in many complex lesions and the operative mortality is low, its success must be judged by the state of long-term survivors.

Though calcification has occurred within a year in 5 of the 7 patients, it does not appear to have influenced function of the homograft valve. There has been no evidence of pro-

gressive stenosis and pulmonary valve closure remains clear without increased delay which suggests the valve is pliable and not narrowed. Nor does our crude method of assessing pulmonary regurgitation suggest that this is increasing. Calcification occurred as rapidly in the adults as in the children and adolescents in these series. We cannot explain why in 2 children it has not been seen even with careful screening 2 to 3 years after operation. The graft used in one had been ethylene oxide sterilized after being in streptomycin and penicillin for 6 days and then subsequently gamma-irradiated. The only comment about this valve was by the technician who noted it was a 'most beautiful valve' as was the valve used in Case 7 which had been gamma-irradiated. All the valves except in Case 7 had been in antibiotic solution, and it was impossible to find any preparation factors which differed in those that calcified and those that did not. The calcification in the right-sided grafts involves the aortic wall and not the cusps over a 5-year period, whereas in the aortic grafts in the aortic position the calcification may be exuberant on the valve cusps but occurs less commonly. The whole problem of the pattern of calcification in these various grafts

remains unsolved. Though it is tempting to postulate that calcification in the right-sided grafts is related to venous blood content or right-sided haemodynamics, this cannot be the explanation because of the rarity of calcification in aortic homografts put in the pulmonary position (J. Somerville, unpublished data) when pulmonary autografts are used as aortic valve substitutes (Ross, 1967). It cannot alone be related to differing calcium metabolism of childhood since it appeared quicker in two adults. Possibly there is a different metabolism of ionized calcium in cyanotic patients and children. Whatever the cause of the relatively rapid calcification of the aortic tube, it does not appear to alter function of the valve cusps which remain mobile. The valve opens freely and shows no evidence of progressive shrinkage or rupture even in the case with persistent pulmonary hypertension.

It has been suggested that replacement and reconstruction of the right ventricular outflow in extreme Fallot and pulmonary valve atresia is an unnecessary refinement adding a further mortality to radical corrective surgery. We do not believe that it is satisfactory in the long term to leave severe pulmonary regurgitation in a patient with a diseased hypertrophied right ventricle and persistent right heart failure after conventional repair of simple type I pulmonary atresia.

Providing confirmation of this view has been the need to put in homografts in the pulmonary position in two patients operated on in other centres for pulmonary valve atresia and Fallot with extreme outflow hypoplasia, who were disabled by persistent right heart failure. For type 2 or 3 pulmonary atresia there is no alternative but to use a graft. Including these more difficult lesions we accept a slightly higher mortality of 12 per cent for this radical correction, comparing favourably with the lower mortality of 8 per cent for conventional Fallot repair of the simpler lesions. This increased mortality is related mainly to haemorrhage and renal complications that inevitably arise from a long difficult bypass.

The aortic homograft has not only been useful in both pulmonary atresia and Fallot, but the technique has been successfully modified for use in truncus (Rastelli, Titus, and McGoon, 1967; Wallace *et al.*, 1969), transposition with pulmonary stenosis (Rastelli, Wallace, and Ongley, 1969), severe pulmonary stenosis (Marchand, 1967), and 'Fallot' with absent or rudimentary pulmonary valves (Ross and Somerville, in preparation). Rapid calcification has also been

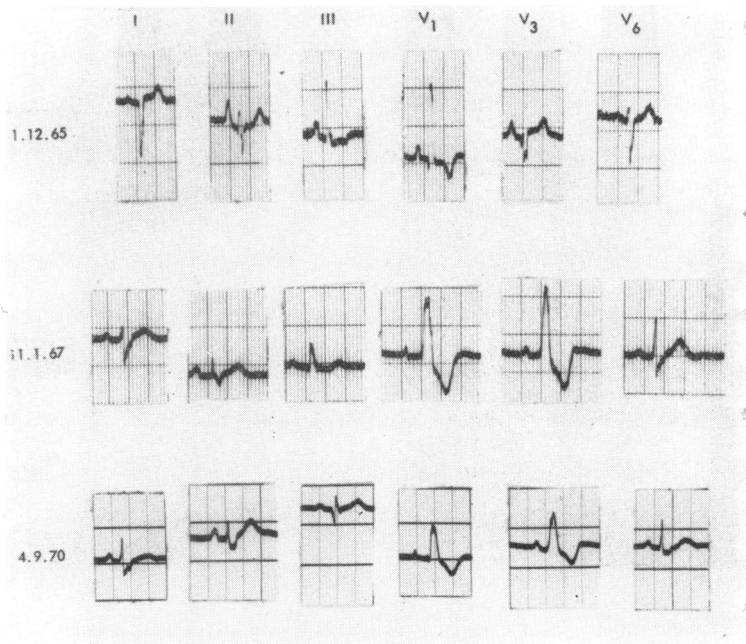
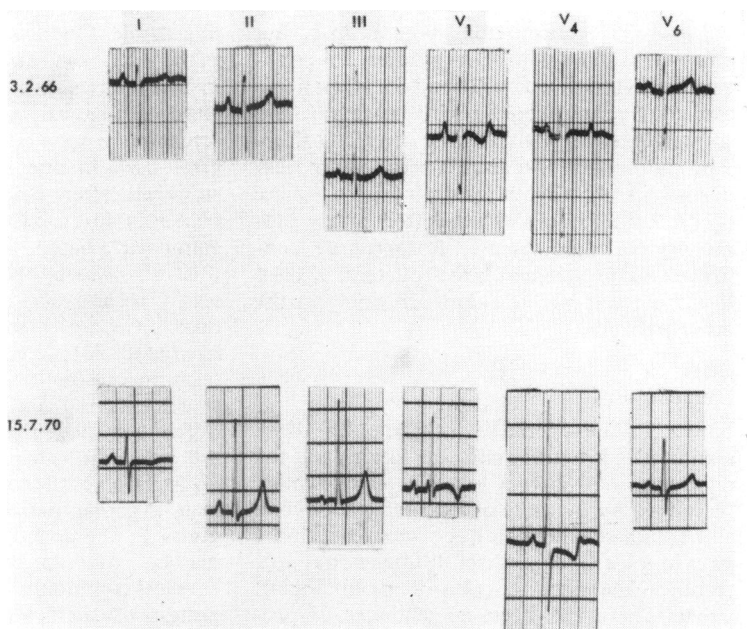


FIG. 5 Electrocardiogram from Case 2 showing complete right bundle-branch block which developed immediately after operation. In 1970 there is diminution in the right ventricular R waves and return of the axis towards normal.

FIG. 6 Electrocardiogram before and 4½ years after operation in Case 1 showing increase in right ventricular hypertrophy and diminution of right atrial hypertrophy.



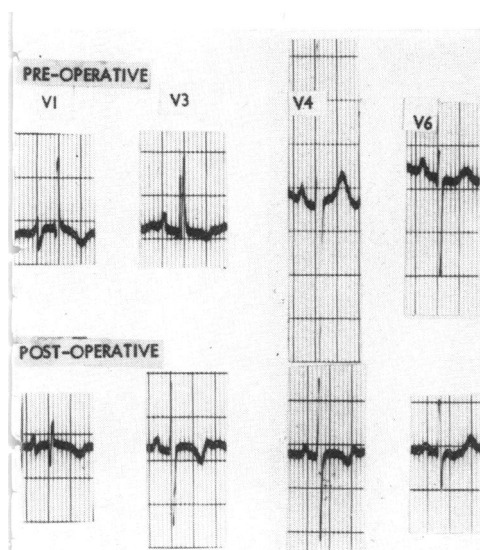


FIG. 7 Electrocardiogram from Case 3 before and 2 years after operation showing incomplete regression of right ventricular hypertrophy.

noted in truncus repair, but the continued good function of these grafts in pulmonary atresia and Fallot over a longer period of years justified continued use of this technique for these and other lesions.

Fascia lata reconstruction of the outflow has been tried in Fallot and pulmonary atresia, but in our experience this has proved to be disastrous owing to progressive shrinkage of the graft after 6 to 12 months (Ross and Somerville, 1971). Currently, fresh homografts stored in antibiotic solution for less than 2 weeks are being used and it is hoped

this will prolong the life and function of the graft.

One of the major problems in treating pulmonary atresia is to avoid the problems of severe pulmonary vascular disease as was present in the first patient. Whether the pulmonary hypertension is related to left heart failure suggested by the pulmonary oedema and high wedge pressure which subsequently fell, or whether there was true pulmonary vascular disease is not known. The latter is likely as there were 5 large collaterals perfusing the right lung and the child had been mildly cyanotic with many respiratory infections in infancy, which suggested a large pulmonary blood flow at some time. Patients with this history are only considered for operation after most careful study, particularly if there is fixed anterior symmetrical bulging of the chest, suggesting altered lung compliance. Greater efforts are now made to measure the pulmonary artery pressure before operation in such patients, and where possible at an earlier age we may recommend elective thoracotomy to ligate collaterals and create a shunt into a pulmonary artery in patients where true central pulmonary arteries are demonstrated to be present. Though this may not improve the patient's arterial oxygen saturation at the time, it is hoped that it may prevent irreversible pulmonary vascular disease, help develop the true pulmonary artery, and make the subsequent radical correction easier as these large collaterals lie posteriorly and are inaccessible through the median sternotomy at the time of definitive correction.

Seven of the first nine patients had previous shunt operations. Not only is this often necessary for survival to an age where more radical surgical correction is possible but also an elective shunt is favoured in patients where

TABLE 2 Postoperative haemodynamic data in patients with homograft reconstruction of right ventricular outflow tract

| Case No. | Time after op. | Pressures (mmHg) | | | | Saturation O ₂ % | | | Art. |
|----------|----------------|------------------|--------|----------|--------|-----------------------------|----|-------|------|
| | | PCV | PAP | RV | Syst. | PA | RV | RA | |
| 1 | 1 mth | 30 | 65/35 | 90/6 | 80/50 | 70, 79 | 68 | 68 | 93 |
| | 1 yr | 20/13 | 75/12 | 100/5 | 110/50 | 69 | 66 | 68 | 97 |
| | 4½ yr | | 85/35 | 140/5-12 | 140/75 | 74 | 66 | 62-71 | 93 |
| | 4½ yr | | 135/45 | 190/8 | | | 51 | | 94 |
| 2 | Refused | — | — | — | — | — | — | — | — |
| 3 | 7 mth | | 15/5 | 47/0 | 80/40 | 68 | 66 | 62 | 100 |
| | 4 yr | | 15/5 | 35/5 | 110/70 | 70 | 69 | 72 | 99 |
| 4 | 3 yr | 15/13 | 38/10 | 63/5 | 130/70 | 78 | 69 | 69 | 92.5 |
| 5 | 10 mth | | 24/10 | 45/0.5 | 85/65 | 70 | 68 | 71 | 95 |
| 6 | 10 mth | | 14/8 | 65/10.5 | 135/85 | 76 | 76 | 76 | 98 |
| | 3 yr | | 25/5 | 45/7 | 110/60 | 74 | 73 | 76 | 98 |
| 7 | 1 yr | | 20/8 | 40/0.6 | 115/65 | 76 | 74 | 75 | 96 |

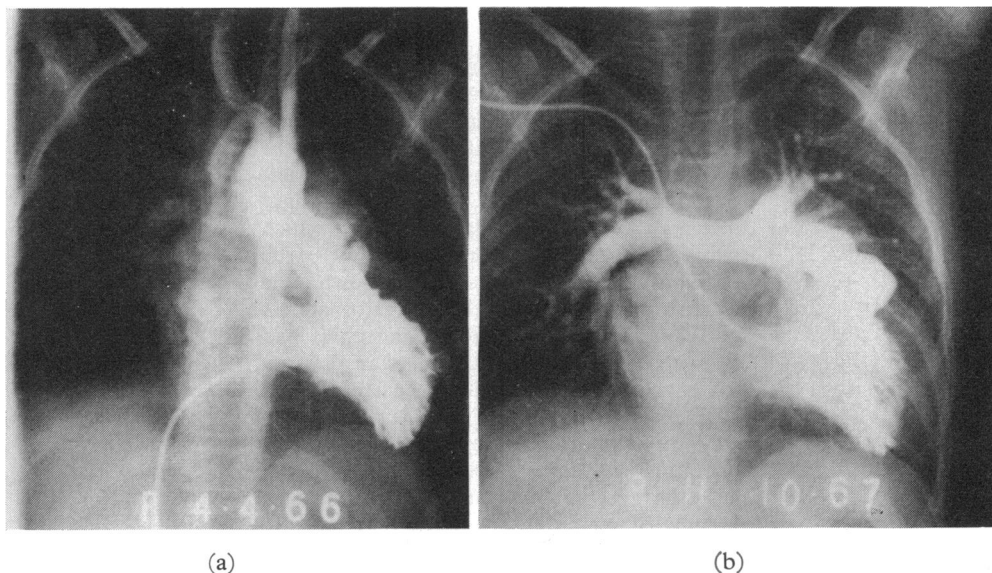


FIG. 8 Right ventricular angiocardigram before and after total correction in Case 3.
 (a) Pulmonary valve atresia shown with early opacification of right Blalock anastomosis.
 (b) Investigation 7 months after operation showing bulged right ventricular outflow, which has not increased in size, and normal pulmonary vascular pattern.

the haemoglobin is above 18 g or the child is under 8 years, in order to reduce the haemoglobin and develop the left side of the heart and thus minimize the bleeding and pulmonary oedema problems. We favour early shunting of these patients to prevent thrombotic complications both in the lungs and systemic circulation and in order to encourage development of the central pulmonary arteries. In following these patients before operation we are guided by the height of the haemoglobin in choosing the time for an elective shunt operation.

The shunt of choice is Waterston's anastomosis as it is easy to close at the time of radical correction (Somerville *et al.*, 1969). Care is required to prevent upward kinking of this anastomosis in the infant, as if this occurs resulting in sole perfusion of the right lung there may be inadequate development of the pulmonary arteries, thus making total correction in a type 1 or 2 pulmonary atresia ultimately inoperable.

Those patients living on a sizeable duct may not require this before total correction. Except under unusual circumstances, radical correction with the use of a homograft would not be done electively under the age of 8 years because of the difficulties of fitting an adult graft. It is considered that the long-term well-being of patients who have had repair of pulmonary atresia and extreme

Fallot using a homograft have justified this operation.

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